Visual Vignette

Idiopathic Intracranial Hypertension: A unique etiology of pseudomeningocele and resultant empty sella and CSF rhinorrhea

Case Presentation: A 41-year-old female presented with a four-month history of right-sided nasal congestion and copious clear drainage. Her symptoms were attributed to seasonal allergies and treated with intranasal glucocorticoids and decongestants. Despite medical therapy, her symptoms persisted and a CT scan of the sinuses was obtained. Imaging demonstrated a fluid-attenuating lesion extending through the sellar region and into the right sphenoid sinus and nasopharynx via a skull base osseous defect (Fig. 1A and 1B). Beta-2 transferrin testing of her rhinorrhea sample was positive, reflecting a cerebrospinal fluid (CSF) leak. MRI of the brain confirmed the presence of a pseudomeningocele advancing through the sellar region into the right nasal cavity (Fig. 1C), with resultant partially empty sella, flattening of the posterior sclera, and prominent fluid-filled optic nerve sheaths. No clinical or biochemical evidence of pituitary dysfunction was noted on further evaluation. Lumbar puncture was performed and notable for an opening pressure of 34 cm of water. What is the diagnosis?

Answer: The patient was diagnosed with idiopathic intracranial hypertension (IIH). The uncommon presentation of CSF rhinorrhea was secondary to pseudomeningocele protrusion through a skull base defect in the sphenoid sinus and into the nasal cavity. The active CSF leak was postulated to provide partial relief of her increased intracranial pressure, leading to the lack of classical IIH symptoms, such as headache, visual disturbances, tinnitus, or diplopia (1,2). Additional imaging findings, which supported a diagnosis of IIH, included flattening of the posterior globe, prominent optic nerve sheath fluid and empty sella (3). The importance of recognizing a pseudomeningocele-related CSF leak, with associated partially empty sella, is critical, as this potentially dangerous entity warrants evaluation by endocrinology and neurosurgery. The patient underwent pseudomeningocele resection and CSF leak repair via a transsphenoidal approach and subsequently acetazolamide therapy was initiated for treatment of IIH. A combination of persistent rhinorrhea with pseudomeningocele and empty sella on imaging should raise clinical suspicion for underlying IIH.

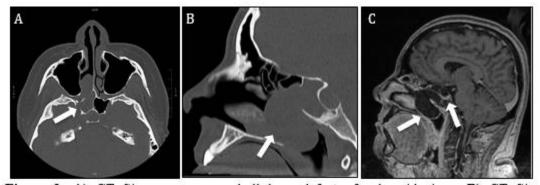


Figure 1. A) CT Sinuses: osseous skull base defect of sphenoid sinus. B) CT Sinuses: pseudomeningocele extending through the sphenoid sinus. C) MRI brain: partially empty sella and pseudomeningocele within the nasopharynx.

Disclosure

The authors have no multiplicity of interest to disclose.

The views expressed are those of the authors and do not reflect the official views or policy of the Department of Defense or its Components.

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